

# Incidental diagnosis of Coronary Anomaly of the Circumflex Artery with Retroaortic Course by Transthoracic Echocardiography in young Athlete

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**Received Date:** February 07, 2024; **Accepted Date:** February 14, 2024; **Published Date:** February 22, 2024.

**Citation:** Massimo Bolognesi, (2024), Incidental diagnosis of Coronary Anomaly of the Circumflex Artery with Retroaortic Course by Transthoracic Echocardiography in young Athlete, *Clinical Research and Clinical Reports*, 4(1); **DOI:**10.31579/2835-8325/065

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## Abstract

The morphology of human coronary anatomy is fascinating because it is variable. When the morphology deviates from typical anatomy, it is considered anomalous, but this occurs in less than 1% of the population. The left circumflex anomaly is one of the most common congenital anomalies of the coronary arteries, whether it originates from the right coronary sinus (RCS), ergo the right sinus of Valsalva (RSV), or from the proximal right coronary artery and cusp. In this report, the author describes the original case of a 22-year-old healthy male and fit athlete with an anomalous course and origin of the LCX, diagnosed on TTE by the typical RAC during sports pre-participation screening.

**Key words:** congenital anomalies of coronary artery; rac sign; echocardiography; computed coronary tomography angiography; athlete

## Introduction

The morphology of human coronary anatomy is fascinating because it is variable. When the morphology deviates from typical anatomy, it is considered anomalous; however, this occurs in less than 1% of the population [1]. In a normal coronary tree, the left circumflex artery (LCX) arises from the left main artery, which in turn arises from the left coronary sinus of Valsalva [2]. One or two obtuse marginal branches arise from the LCX after entering the atrioventricular groove (AVG) [1]. The left circumflex anomaly is in fact one of the most common congenital anomalies of the coronary arteries, whether it originates from the right coronary sinus (RCS), ergo from the right sinus of Valsalva (RSV), or from the proximal right coronary artery and cusp [3]. It should be noted that the retroaortic pathway of an artery originates from the posterior course between the interatrial septum and the non-coronary sinus. This anomalous pathway also occurs in various forms of congenital coronary artery disease, such as the ectopic origin of the left main and LCX arteries [4]. In practice, this congenital anomaly is documented by both coronary computed tomography angiography (CCTA) and invasive coronary angiography (ICA) [5]. In addition, Witt and coworkers shed light on the relationship between abnormal coronary arteries and the retroaortic coronary sign (RAC) on transthoracic echocardiography (TTE) [3]. Indeed, the RAC sign can be seen in the apical 4-chamber view as a tubular image above the mitral valve plane, directed towards the right sinus of Valsalva [6]. However, this can be confused with mitral annular calcification, calcified valves or even normal coronary arteries [3]. Other peculiar echocardiographic signs have been noted in the parasternal long-axis view of TTE: "Bleb's sign", which is an additional round structure below the non-coronary cusp sign [6]. Usually, the

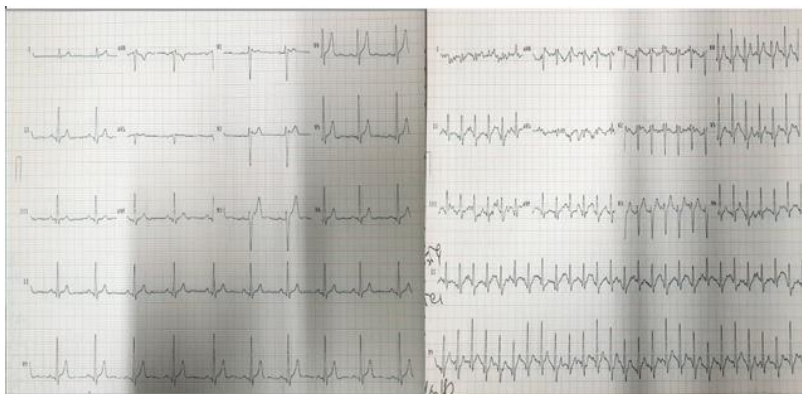
retroaortic course of an anomalous LCX artery is considered benign if it has no haemodynamic significance for ischaemic heart disease [4]. In this report, the author describes the original case of a 22-year-old healthy male and fit athlete with an anomalous course and origin of the LCX, diagnosed on TTE by the typical RAC ultrasound sign; examination performed as a pre-participation screening and subsequently confirmed by coronary computed tomography angiography (CCTA).

## Case Report

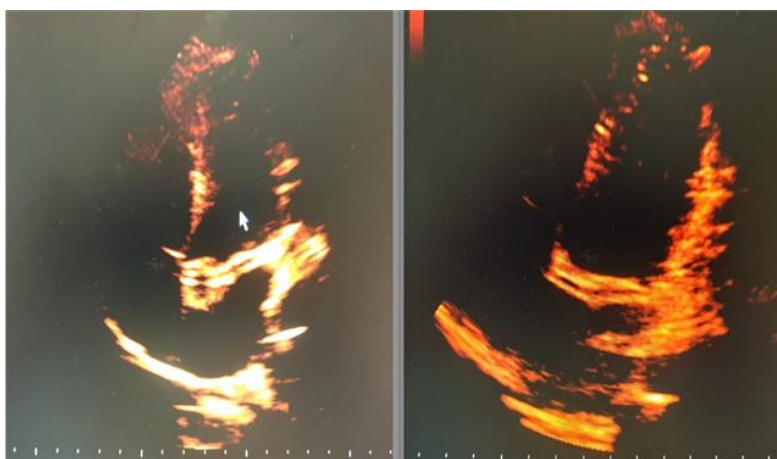
A healthy and fit 22-year-old male athlete presenting to our sports cardiology centre underwent routine cardiac assessment as part of his pre-participation screening. His medical history was unremarkable. His physical examination was within normal limits. The resting ECG (Fig. 1) was normal, and the exercise test on the cycloergometer was included in the ST ECG monitoring and was effective, reaching 100% of the age-predicted maximum heart rate. The athlete showed no clinical symptoms on exercise and demonstrated excellent functional capacity, adequate blood pressure behaviour and, in particular, the absence of arrhythmias or electrocardiographic criteria for myocardial ischaemia. 2D transthoracic echocardiography showed no ventricular dilatation with preserved global and segmental contractility; left ventricular ejection fraction (LVEF) and global longitudinal strain (GLS) were within the normal range. Diastolic function parameters were preserved and the heart valves showed no significant anatomical or functional changes. However, a hyperechoic tubular image (characteristic of retroaortic course) was observed in the apical windows (Figure. 2), suggesting an anomalous retroaortic coronary course (RAC) of the LCX artery. Therefore, an

anomalous congenital left CX coronary artery was suspected. For this incidental echocardiographic finding, coronary computed tomography angiography (CCTA) was requested and added to the diagnosis, confirming the observed echocardiographic sign. The 3D image reconstruction (Figure. 3) showed a retroaortic course of the left circumflex artery originating from

the right coronary sinus in a single ostium and at an acute angle ( $30^\circ$ ), without evidence of intramural course or extrinsic compression by the aortic root. On the basis of this evidence, the athlete was deemed fit to participate in sport competition.



**Figure 1: Electrocardiographic tracing of the exercise stress test (left at rest and right at peak exercise) without electrocardiographic evidence of myocardial ischaemia on exercise.**



**Figure 2: Five-chamber (on the left) and Subcostal (on the right) echocardiogram views showing the RAC sign (white arrow), characteristic of an abnormal LCX Artery with a retroaortic course.**



**Figure 3: 3D reconstruction of CT coronary angiography showing the coronary anatomy with an anomalous LCxA originating from a single ostium and its retroaortic course.**

## Discussion and Conclusion

The RAC sign is a new echocardiographic feature that describes the often-occasional finding of an anomalous coronary artery. It is typically associated with abnormalities of the left coronary artery, particularly the left circumflex artery. Few echocardiographic signs correlate with this feature. It is often underdiagnosed on transthoracic echocardiograms because of confusion with artefacts, calcifications and other cardiac structures. The Italian COCIS 2023 protocol makes the following recommendations for granting eligibility to individuals with coronary artery anomalies. In young and very young athletes with anomalous origin of the left circumflex artery from the right sinus or from the right coronary artery (with retroaortic course), a "liberal" attitude is justified, but based on a careful study with CCTA of the abnormal coronary artery and the documented absence of maximal exercise ischaemia; in such a case, with appropriate periodic follow-up, eligibility can be granted for all sports. In elite and master athletes with this anomaly, the assessment of fitness must be weighted more heavily because of the possible association with atherosclerotic coronary artery disease. It is therefore of the utmost importance that cardiologists and sports physicians are aware of the RAC sign and its significance, as it may provide important clues to a correct diagnosis. It should be recognised that the RAC sign may mimic other common TTE findings, such as image artefacts and annular or valvular calcifications, and should therefore be carefully differentiated from these. This case confirms the importance of transthoracic echocardiography as a non-invasive tool for the diagnosis of coronary artery abnormalities, particularly in the context of sports preparticipation screening. In fact, these anomalies are usually diagnosed by coronary computed tomography

angiography and coronary angiography, especially in the presence of the echocardiographic finding of retro aortic coronary sign or "crossed aorta sign".

## Conflict of Interest

The author has no financial conflicts of interest

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